AMENDMENTS TO THE CLAIMS

Docket No.: A0345.0026

1. (Original) A compound of formula (I) or a pharmaceutically acceptable salt or prodrug thereof:

HO
$$\stackrel{\mathsf{R}}{\longrightarrow}$$
 $\stackrel{\mathsf{N}}{\longrightarrow}$ $\stackrel{\mathsf{N}}{\longrightarrow}$

wherein

R is C₁₋₃ alkylAr¹ where Ar¹ is phenyl or pyridyl;

wherein phenyl is substituted by one or more substituents selected from CN, CON(R¹)₂, SO_nR², SO₂N(R¹)₂, N(R⁵)₂, N(R¹)COR², N(R¹)SO_nR², C₀₋₆ alkylAr², C₂₋₆ alkenylAr² and C₃₋₆ alkynylAr² wherein one or more of the -CH₂- groups of the alkyl chain may be replaced with a heteroatom selected from O, S and NR³, provided that when the heteroatom is O, at least two -CH₂- groups separate it from any additional O atom in the alkyl chain; or two adjacent substituents on the Ar¹ phenyl may together form a fused 5- or 6-membered saturated or unsaturated ring wherein the ring optionally contains 1 or 2 heteroatoms selected from O, S and NR⁴ and is optionally substituted by one or more substituents selected from, an oxo group, C₁₋₆ alkyl and C₀₋₃ alkylAr⁴;

and the Ar¹ phenyl is optionally substituted by one or more additional substituents selected from F, Cl, Br, CF₃, OCF₃, OR³ and C₁₋₆ alkyl;

and wherein pyridyl is substituted by one or more substituents, selected from, CN, $CON(R^1)_2$, SO_nR^2 , $SO_2N(R^1)_2$, $N(R^5)_2$, $N(R^1)COR^2$, $N(R^1)SO_nR^2$, F, Cl, Br, CF₃, OCF₃, OR³, C₁₋₆ alkyl, C₀₋₆ alkylAr², C₂₋₆ alkenylAr² and C₃₋₆ alkynylAr² wherein one of the -CH₂- groups of the alkyl chain may be replaced with a heteroatom selected from O, S and NR^3 , provided that when the heteroatom is O, at least two -CH₂- groups separate it from any additional O atom in the alkyl chain; or two adjacent substituents on the Ar¹ pyridyl may together form a fused 5- or 6-membered saturated or unsaturated ring wherein the ring optionally contains 1 or 2 heteroatoms selected from O, S and NR^4

Docket No.: A0345.0026

and is optionally substituted by one or more substituents selected from, an oxo group, C_{1-6} alkyl and C_{0-3} alkyl Ar^4 ;

 R^1 is H, C_{1-6} alkyl optionally substituted by OH, Ar^3 , or C_{1-6} alkyl Ar^3 , or the group $N(R^1)_2$ may form a 5- to 10-membered heterocyclic group optionally containing one or more additional heteroatoms selected from O, S and NR^3 and is optionally substituted by an oxo group;

 R^2 is C_{1-6} alkyl optionally substituted by OH, Ar^3 , or C_{1-6} alkyl Ar^3 ;

R³ is H, or C₁₋₆ alkyl;

 R^4 is H, C_{1-6} alkyl or C_{0-3} alkyl Ar^4 ;

 R^5 is H, C_{1-6} alkyl optionally substituted by OH, Ar^3 , or C_{1-6} alkyl Ar^3 , or the group $N(R^5)_2$ may form a 5- to 10-membered heterocylic group optionally containing one or more additional heteroatoms selected from O, S and NR^3 and is optionally substituted by an oxo group;

 Ar^2 and Ar^3 are independently phenyl or a 5- to 10-membered heteroaryl group containing up to 3 heteroatoms selected from O, S and NR³, which may be optionally substituted by one or more substituents selected from F, Cl, Br, CN, CF₃, OCF₃, OR³ and C₁₋₆ alkyl;

Ar⁴ is phenyl or pyridyl either of which may be optionally substituted by one or more substituents selected from F, Cl, Br, CN, CF₃, OCF₃, OR³ and C₁₋₆ alkyl; and

n = 0, 1 or 2;

provided that the compound is not:

- a) 3,4,5-piperidinetriol, 1-[(1,1'-biphenyl)-4-ylmethyl]-2-(hydroxymethyl)-, (2R,3S,4R,5S);
- b) 3,4,5-piperidinetriol, 2-(hydroxymethyl)-1-[(4-methoxyphenyl)methyl]-, (2R,3S,4R,5S);
- c) 3,4,5-piperidinetriol, 2-(hydroxymethyl)-1-[(4-methylthiophenyl)methyl]-, (2R,3S,4R,5S);

- d) acetamide, N-[4-[[3,4,5-trihydroxy-2-(hydroxymethyl)-1-piperidinyl]methyl]phenyl]-, (2R,3S,4R,5S); or
- e) 3,4,5-piperidinetriol, 2-(hydroxymethyl)-1-[(4-methoxy-3-methylphenyl)methyl]-, (2R,3S,4R,5S).
 - 2. (Original) A compound as defined in claim 1 wherein R is C₁ alkylAr¹.

- 3. (Currently amended) A compound as defined in claim 1 [[or 2]] wherein Ar¹ is phenyl, wherein phenyl is substituted as defined for claim 1.
- 4. (Currently amended) A compound as defined in any one of the preceding elaims claim 1, wherein Ar¹ is phenyl, wherein phenyl is substituted by one or more substituents selected from CN, CON(R¹)₂, N(R⁵)₂, and C₀₋₆ alkylAr², wherein one or more of the -CH₂- groups of the alkyl chain may be replaced with a heteroatom selected from O, S and NR³, provided that when the heteroatom is O, at least two -CH₂- groups separate it from any additional O atom in the alkyl chain, or two adjacent substituents on the Ar¹ pyridyl may together form a fused 5- or 6-membered saturated or unsaturated ring, wherein the ring optionally contains 1 or 2 heteroatoms selected from O and NR⁴ and is optionally substituted by one or more substituents selected from, an oxo group, C₁₋₆ alkyl and C₀₋₃ alkylAr⁴[[,]]; and the Ar¹ phenyl is optionally substituted by one or more additional substituents selected from F, Cl, Br, CF₃, OCF₃, OR³ and C₁₋₆ alkyl.
- 5. (Currently amended) A compound as defined in any one of the preceding elaims claim 1, wherein Ar¹ is phenyl, wherein phenyl is substituted by one or more substituents selected from CN, CON(R¹)₂, N(R⁵)₂, and C₀₋₆ alkylAr², wherein one or more of the -CH₂- groups of the alkyl chain may be replaced with O, provided that at least two -CH₂- groups separate it from any additional O atom introduced into the alkyl chain; and the Ar¹ phenyl is optionally substituted by one or more additional substituents selected from F, Cl, Br, CF₃, OCF₃, OR³ and C₁₋₆ alkyl.

6. (Currently amended) A compound as defined in any one of the preceding claims claim 1, wherein Ar^2 is phenyl which is optionally substituted by one or more substituents selected from F, Cl, Br, CN, CF_3 , OCF_3 , OR^3 and C_{1-6} alkyl.

- 7. (Currently amended) A compound as defined in any one of the preceding claims claim 1, wherein R¹ is H or C₁₋₆ alkylAr^{3.}
- 8. (Currently amended) A compound as defined in any one of the preceding claims claim 1, wherein R^4 is H or C_{1-6} alkyl.
- 9. (Currently amended) A compound as defined in any one of the preceding claims claim 1, wherein Ar^3 is phenyl which may be optionally substituted by one or more substituents selected from F, Cl, Br, CN, CF_3 , OCF_3 , OR^3 and C_{1-6} alkyl.
- 10. (Currently amended) A compound as defined in any one of the preceding claims claim 1, wherein R^5 is C_{1-6} alkyl.
- 11. (Original) A compound of formula (I) as described in Example 1 or a pharmaceutically acceptable salt or prodrug thereof.
 - 12. (Cancelled).
- 13. (Currently amended) A pharmaceutical composition comprising a compound of formula (I) as defined in any one of claims 1 to 11 claim 1, together with one or more pharmaceutically acceptable carriers, excipients and/or diluents.
- 14. (Currently amended) A process for the preparation of a compound of formula (I) as defined in any one of the preceding claims claim 1, which process comprises:
- a) conducting reductive amination of an aldehyde of formula R^6CHO_1 wherein R^6 is C_{0-2} alkyl Ar^1 where Ar^1 is as defined in claim 1 with 1-deoxygalactonojirimycin [2-(hydroxymethyl)-3,4,5-piperidinetriol, (2R,3S,4R,5S)] (II):

Docket No.: A0345.0026

- b) alkylation of alkylating 1-deoxygalactonojirimycin (II) with an activated species R⁶CH₂X, wherein R⁶ is as defined above and X is a leaving group; or
- c) *N*-acylation of *N*-acylating a protected derivative of 1-deoxygalactonojirimycin (II) with an activated acyl derivative, followed by reduction of the resultant amide with a reducing agent and deprotection.

15-29. (Cancelled).

30. (Original) A compound of formula (III):

wherein R is as defined in claim 1 and P, which may be the same or different, are hydroxy protecting groups.

- 31. (New) A method for preventing or treating a glycolipid storage disease, comprising administering to a subject in need thereof the compound of claim 1.
- 32. (New) The method of claim 31, wherein said glycolipid storage disease is Gaucher disease, Sandhoffs disease, Tay-Sachs disease, Fabry disease or GM1 gangliosidosis.
- 33. (New) A method for preventing or treating a disorder or disease associated with abnormal glycolipid synthesis, comprising administering to a subject in need thereof the compound of claim 1.

34. (New) The method of claim 33, wherein said disorder or disease is cancer.

- 35. (New) The method of 34, wherein said cancer is selected from the group consisting of brain cancer, neuronal cancer, neuroblastoma, renal adenocarcinoma, malignant melanoma, multiple myeloma and multi-drug resistant cancer.
- 36. (New) A method for preventing or treating a disorder or disease associated with abnormal glycolipid metabolism, comprising administering to a subject in need thereof the compound of claim 1.
- 37. (New) The method of claim 36, wherein said disorder or disease is Niemann-Pick disease type C, mucopolysaccharidosis type I, mucopolysaccharidosis type IIIA, mucopolysaccharidosis type IIIB, mucopolysaccharidosis type VI, mucopolysaccharidosis type VII, α-mannosidosis or mucolipidosis type IV.
- 38. (New) The method of claim 36, wherein said disorder or disease is selected from the group consisting of Alzheimer's disease, epiliepsy, stroke, Parkinson's disease and spinal injury.
- 39. (New) A method for preventing or treating an infectious disease caused by an infectious microorganism that utilizes glycolipids on the surface of cells as receptors for either said microorganism itself or for toxins produced by said microorganism, or by an infectious microorganism that requires the synthesis of glucosylceramide to sustain an infection, said method comprising administering to a subject in need thereof the compound of claim 1.
- 40. (New) A method for preventing or treating a condition treatable by the administration of a ganglioside, said method comprising co-administering to a subject in need thereof said ganglioside and the compound of claim 1.

41. (New) The method of claim 40, wherein said ganglioside is a GM1 ganglioside.

- 42. (New) A method for reversibly rendering a male mammal infertile, comprising administering to said mammal the compound of claim 1.
- 43. (New) A method for treating obesity, comprising administering to a subject in need thereof the compound of claim 1.

- 44. (New) A method for treating an inflammatory disorder or disease associated with macrophage recruitment and activation, comprising administering to a subject in need thereof the compound of claim 1.
- 45. (New) The method of claim 44, wherein said inflammatory disorder or disease is selected from the group consisting of rheumatoid arthritis, Crohn's disease, asthma and sepsis.